AUDIOLOGIC CONSEQUENCES OF CRANIOFACIAL ABNORMALITIES AND THE ROLE OF THE AUDIOLOGIST ON THE CRANIOFACIAL INTERDISCIPLINARY TEAM

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ABSTRACT

Craniofacial abnormalities refer to malformations or anomalies of the face or head. When a child is born with a craniofacial abnormality, various aspects of growth and development can be affected. Among these are malformations and dysfunctions of the auditory system. Among children with both syndromic and non-syndromic craniofacial diagnoses, various forms of hearing loss are present. Due to these congenital malformations, this patient population is at a higher risk for conductive, sensorineural and mixed hearing losses. For this reason, it is imperative that the auditory system of these children be monitored closely by an audiologist as part of a larger craniofacial interdisciplinary pediatric team.

DEDICATION

This capstone paper is dedicated to my parents, Jim and Peggy Ruwe. Without their support and assistance, I would not have had the opportunity to successfully complete this capstone paper or the Au.D. program. I would also like to dedicate this to the memory of my grandfather, Joseph Ruwe, who provided me an excellent example of hard work and dedication.

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CHAPTER 1

INTRODUCTION

Craniofacial abnormalities define a class of congenital disorders that affect growth and development of the skull and facial bones. These abnormalities can affect various aspects of a developing child's life: medical, social, developmental and educational. Due to the physical characteristics of these disorders, they are usually diagnosed at or close to birth. Variations of craniofacial abnormalities can range from mild to severe deformities and can affect various aspects of growth and development of the child. Anomalies that commonly arise from craniofacial defects include: cleft lip/cleft palate, craniosynotosis and hemifacial microsomia. For many children, craniofacial abnormalities are secondary to a congenital syndrome such as Treacher-Collins, Pierre-Robin, Pfeiffer, Down and Apert Syndromes. However, for other children, craniofacial abnormalities are caused by single mutations and are not associated with a syndrome.

No one single factor is associated with causing all craniofacial abnormalities. As with many abnormalities of congenital origin, various factors can contribute to prenatal development, ultimately affecting the development of cranial structures. Genetic makeup may be one cause of craniofacial abnormalities. Specific syndromes such as Turner, Down and Goldenhar's Syndromes are linked to specific mutations, deletions or additions in the child's genetic map. Environmental factors during prenatal development may also be associated with craniofacial abnormalities. One example is Fetal Alcohol Syndrome

(FAS) from which the baby's prenatal maternal exposure to alcohol can cause specific craniofacial characteristics, including malformations in the development of the skull. Finally, research is being conducted on prenatal nutritional deficiencies and their correlation to the development of craniofacial abnormalities. One example of this is folic acid deficiency, which has been linked to cleft lip and/or cleft palate (Bianchi, Calzolari, Ciulli, Cordier, Gualandi, Pierini & Mossey, 2000). Specific postnatal genetic testing can be performed on the child and the parents to help to find the specific cause of the craniofacial abnormality.

A significant relationship between craniofacial abnormalities and hearing loss is recognized in children. It is estimated that 82% of the 71 syndromes and birth anomalies associated with hearing loss are classified as craniofacial abnormalities (Jones, 1988). For this reason, it is important for an audiologist to understand the characteristics of craniofacial abnormalities and associated hearing losses. This review will describe the impact various craniofacial abnormalities can have on the auditory system and the role an audiologist must take in the proper diagnosis and management of hearing loss for this population as part of an interdisciplinary craniofacial team.

CHAPTER II

AUDIOLOGIC ASPECTS OF CRANIOFACIAL ABNORMALITIES

Auditory Pathways Abnormalities

The developing auditory system requires proper anatomical structure formation and function to efficiently send information to the auditory cortex. For many children with craniofacial abnormalities, structural malformations in the outer and middle ear can cause significant degrees of conductive hearing losses. Inner ear deformities such as irregularities of the cochlea and vestibular system can result in sensorineural hearing losses and balance-related symptoms. Because of the negative impact a hearing loss can have on speech, language and overall development, children with craniofacial abnormalities should be closely evaluated and monitored by an audiologist for early identification and intervention (Maddell, 2008).

Outer ear malformations can include microtia, a small or underdeveloped pinna, anotia, the complete absence of a pinna, and atresia, a small or absent external ear canal. These malformations can cause a range of effects on hearing, from no noted difficulties to a maximal conductive hearing loss. Children can also be born with ear tags or pits and no other associated ear or facial malformations. While a craniofacial team may not follow these children, they should be considered at risk for hearing impairment and should be closely monitored for hearing loss. Roth et al. (2008) studied the prevalence of hearing loss in children with preauricular ear pits and skin tags when compared to typically developing children with no external ear deformity. Their study found that the rate of hearing loss in children increased significantly from 1.5 out of 1000 children with

no ear tags or pits to 8 out of 1000 children born with ear tags or pits. Therefore, these children should be screened by otoacoustic emissions or automated auditory brainstem screenings to evaluate the need for additional testing to rule out hearing loss.

There are particular syndromes that are associated with an increased risk of outer ear malformations. One specific example is Velocardiofacial Syndrome, a genetic condition that results in various craniofacial and physical malformations, including cleft palate, elongated faces and visual impairments (Goldberg, Motzkin, Marion, Scambler & Shprintzen, 2005). Children born with Velocardiofacial Syndrome (VCFS) are at a 70% risk of having auricular malformations, including low set and abnormally rotated pinnas (Reyes, LeBlanc & Bassila, 1999). Because of these anatomical differences, children with VCFS are very susceptible to middle ear pathology. Children with hemifacial microsomia, also referred to as oculoauriculo-vertebral spectrum syndromes (OAVS), can also exhibit significant outer ear abnormalities. Oculoauriculo-vertebral spectrum syndromes define the class of syndromes that are characterized by anomalies of the external ear, hemifacial microsomia, a defect to the development of the lower half of the facial bones, vertebral column abnormalities and are highly associated with overall developmental delay (Bohringer et al., 2005). Children born with OAVS often exhibit ear pits, atresia or stenosis of the external ear canals and malformations of the pinna and middle ear bones. Due to these characteristics, children with OAVS are at a high risk of conductive hearing loss. Rahbar et al. (2001) found that the majority of children with OAVS who had associated hearing loss demonstrated conductive losses (86%). In addition, 10% of children with OAVS also exhibited a sensorineural component.

Therefore, children with syndromes and associated craniofacial abnormalities should be closely evaluated for any type of hearing loss.

Middle ear function is also crucial for proper transduction of sound to the inner ear. Sound delivered to the tympanic membrane by way of the external auditory canal sets the middle ear bones, the ossicles, into mechanical motion. This movement of the middle ear bones delivers the acoustic stimulus present in the auditory canal to the inner ear by way of pressure changes against the oval window. However, when a middle ear pathology is present, this mechanical property of the middle ear function may be disrupted, resulting in a conductive hearing loss.

Because children with craniofacial abnormalities are at a greater risk of having abnormal development of the structures in the middle ear, they also are at a greater risk of exhibiting chronic middle ear pathologies (Di Francesco, Paulucci, Nery & Bento, 2008). The prevalence and consequences of otitis media in this population will be discussed later in this review. Children with middle ear abnormalities can exhibit abnormal development of the tympanic membrane, absent or malformed ossicles, intratymapanic bony masses and closure of the oval window. These all can cause significant conductive hearing loss.

Treacher-Collins Syndrome is a syndrome with a high rate of associated middle ear abnormalities in addition to irregular development of the pinna. Children with Treacher-Collins often demonstrate dsymorphic or absent ossicles on CT scans. While they demonstrate high rate of conductive hearing loss, these children rarely exhibit sensorinerual components to their hearing loss. Pron, Galloway, Armstrong and Posnick (1993) documented 29 individuals with Treacher-Collins and their associated hearing

losses with outer and middle ear abnormalities. Their study documented conductive hearing losses which were mainly attributed to middle ear abnormalities due to malformed or missing ossicles. Another craniofacial syndrome which commonly exhibits these findings is Crouzon's Syndrome, a genetic disorder that typically exhibits abnormalities in the development of the midface, abnormal prominence of the ocular region and craniosynostosis. Children with Crouzon Syndrome also demonstrate significant middle ear anatomical abnormalities due to the presence of craniosynostosis, the early closure of the skull bones (Orvidas, Fabry, Diacova & McDonald, 1999).

In addition to conductive hearing loss in this population, many children with craniofacial abnormalities exhibit significant inner ear abnormalities and resulting sensorineural or mixed hearing losses. Here, abnormalities such as absent or malformed cochleas, vestibular organs, the vestibulocochlear nerve or the auditory processing center of the brain are the cause of the hearing loss. Children with Stickler Syndrome often exhibit sensorineural hearing loss due to incomplete formation of the inner ear. Children born with this diagnosis should be closely monitored for the onset of a sensorineural hearing loss (Nowak, 1998).

Children born with Fetal Alcohol Syndrome (FAS) exhibit characteristic craniofacial features, which aid professionals in the diagnosis of FAS at or close to birth. In addition to other features such as microcephaly, small eyes and thin lips, specific malformations to the auditory system are often apparent. Typically found auditory system abnormalities can include delayed maturation of central nervous system, sensorineural hearing loss and chronic otitis media (Church & Abel, 1998).

Children with craniofacial abnormalities can exhibit malformations and dysfunction of the vestibular system. Cat Scan imaging after birth can assess the integrity of the vestibular system to note possible vestibular disorders in the future. While little research has been conducted on the specific balance disorders in children with craniofacial disorders, the assessment of balance disorders in the pediatric population is beginning to receive more attention (Casselbrant & Mandel, 2005). To better understand the balance system of children with craniofacial abnormalities, full assessments of vestibular, visual and proprioceptive inputs need to be assessed.

As discussed, there are many aspects of the auditory and vestibular system that can be affected when a child is born with a craniofacial abnormality. It is important that the child undergo comprehensive imaging and diagnostic tests to determine the function of the auditory and balance systems. These results will be crucial in determining the most appropriate level of intervention and type of technology for each child.

Cleft Lip/Cleft Palate:

The most common craniofacial anomaly is cleft lip and/or cleft palate. This abnormality is defined by a separation of the bone in the lip and/or the roof of the mouth, the palate. These anomalies, caused by incomplete development during gestation, are identified at birth and are estimated to be found in 1/700 live births (Bianch et al., 2000). Children born with cleft lip and/or cleft palate are at a greater risk of developing difficulties with breathing, feeding, and with speech, language and auditory development (Colburn & Cherry, 1985).

Because of the large number of children born with cleft lip and cleft palate each year, much research has been conducted on the associated hearing loss of these children

and has been well documented for many decades (Gaines, 1940; Gould, 1990; Viswanathan, Vidler, & Richard, 2008). Cleft lip and cleft palate can occur in isolation or as an associated feature of a syndrome. Syndromes such as Stickler and Apert commonly manifest with cleft lip/cleft palate in addition to other characteristics that classify each syndrome.

A child born with a cleft lip and/or cleft palate may also exhibit co-existing abnormalities in the middle ear. For many children, the Eustachian tube, which connects the middle ear space to the nasophyarynx, does not function properly and therefore can result in abnormal function of the middle ear. These abnormalities may cause irregular sound transfer through the middle ear, resulting in a conductive hearing loss. Goudy, Lott, Canady and Smith (2006) investigated the incidence of middle ear pathology and resulting conductive hearing loss in cleft lip/cleft palate children. They found that children born with cleft lip/cleft palate were at a greater risk of conductive hearing loss and required an increased number of myringotomy with pressure equalization (PE) tube surgeries when compared to typically developing children. This procedure involves a tiny incision in the tympanic membrane to remove fluid in the middle ear space and the insertion of a small grommet to assist with equalization between environmental and middle ear pressures (Perkins, 2002).

Phua, Salkeld and de Chalain (2009) and Doyle, Cantekin and Bluestone (1980) also investigated the presence and appropriate management techniques of middle ear pathologies resulting from abnormal development of the middle ear space in cleft lip and cleft palate children. Phau et al. (2009) noted that otolaryngologists are typically more aggressive with the placement of pressure equalization tubes in this population when

compared to typically developing children as they are at a greater risk of recurrent and persistent conductive hearing losses. Currently, there are various philosophies on the degree of aggressiveness of PE tube placement in this population, ranging from proactively placing tubes to using a waiting period of present middle ear pathology as seen in typically developing children. Ponduri, Bradley, Ellis, Brookes, Sandy and Ness (2009) conducted a systematic review of 18 relevant studies that discussed whether routine PE tube insertion in children born with cleft palate was beneficial to hearing and speech and language development when compared to a more conservative approach. They concluded that while some studies found significant benefit of a proactive approach, others reported no increase in these measures and therefore, more evidence is needed in the area to make a definitive clinical recommendation.

Priester and Goorhuis-Brouwer (2008) evaluated the speech and language development of toddler-aged children with and without cleft palate. They found that children with cleft palate were significantly delayed in speech development due to hypernasality. In regards to the effects of hearing loss, they determined that children with properly managed conductive hearing loss exhibited no significant language comprehension or speech articulation delay when compared to children with cleft lip and cleft palate and no associated hearing loss. Because of the possibility of delays in speech and language development, audiologists should closely monitor the hearing of children born with a cleft lip and cleft palate and make recommendations in conjunction with other members of the interdisciplinary craniofacial team.

As the most commonly found craniofacial abnormality, it is important that team members understand how cleft lip and cleft palate affects various aspects of growth and

development. For audiologists and otolaryngologists working with patients who have craniofacial anomalies, associations between cleft lip/cleft palate and middle ear dysfunction should be closely evaluated, monitored and intervened with as appropriate for each child.

Middle Ear Pathology:

Much research has been conducted to evaluate and document the developmental and physical consequences of acute and chronic middle ear pathology in the pediatric population (Brandes & Karsh, 1981; Golz et al., 2005; Roberts et al., 2004). For children with craniofacial abnormalities, the prevalence of middle ear pathology increases when compared to typically developing children (Di Francesco, Paulucci, Nery & Bento, 2008). Because middle ear pathology can have a significant impact on speech and language development, it is important that a child with a craniofacial abnormality be followed closely for middle ear pathology, resulting hearing loss and speech and language delays.

When a child is born with a craniofacial abnormality, the Eustachian tube may not be properly formed or may not function correctly. This, in addition to the physical predisposition any child has to middle ear pathology due to a more horizontal location of the Eustachian tube, can cause the child to have serous otitis media and recurrent middle ear fluid. Because of this common finding, many otolaryngologists are more aggressive with the medical management of middle ear pathology through the use of longstanding PE tubes (CCHMC, 2004).

It has also been documented that proper auditory sensitivity is important for success in the classroom. In a study evaluating the reading abilities of children with a

history of otitis media in comparison with children with no significant history of otitis media, Golz et al. (2005) found that children with recurrent or chronic middle ear pathology in the first five years of life were at a greater risk of delayed reading skills than their peers with no history of middle ear pathology. Because children with craniofacial abnormalities have a higher prevalence of middle ear pathology, educational outcomes should be closely monitored and evaluated for possible delay or risk of future delay.

In addition to significant impact on speech, language and educational development, middle ear pathology can influence other areas of the child's social development. A child's attention and behavior may be questioned as they are more likely to misbehave given a conductive hearing loss than a child who has normal, stable hearing. In a study by Silva, Chalmers and Stewart (1986), teachers reported significantly more behavior problems with children who had history of longstanding bilateral otitis media with effusion when compared to children with no significant history.

Craniofacial abnormalities have been shown to have significant impact on the development and function of each portion of the auditory system. Certain syndromes have associated higher prevalence rates of each type of hearing loss and therefore, a child born with a craniofacial syndrome needs to be monitored for hearing loss if they are at a predisposed higher risk due to their syndrome. These children, particularly children diagnosed with cleft lip and or cleft palate, should be closely monitored by audiology and otolaryngology for middle ear pathology and resulting conductive hearing loss.

CHAPTER III

THE ROLE OF THE AUDIOLOGIST ON THE INTERDISCIPLINARY CRANIOFACIAL TEAM

To help provide appropriate and comprehensive healthcare for a child diagnosed with a craniofacial abnormality, a family may seek the expertise of an interdisciplinary craniofacial team. This team is comprised of various specialists that work in collaboration with one another and the family to make medical, developmental and educational decisions and to provide medical and rehabilitative care to the child. The team follows the child through young adulthood to ensure consistent and comprehensive management of the various needs that may arise during development. Craniofacial teams provide the family with professionals who have experience working with the unique needs of these children.

There are many benefits to seeing a child with a craniofacial abnormality in an interdisciplinary team setting. An interdisciplinary team is a group of professionals that work closely with the family to provide comprehensive medical evaluations and recommendations. Austin et al. (2010) evaluated the access to care of children with craniofacial abnormalities who were seen by an interdisciplinary team and those who saw individual providers. Their research found that children evaluated by a team setting were more likely to receive proper dental care, appropriate auditory evaluations and genetic consultations when compared to children who were seen by individual providers.

Professionals on a craniofacial team are well versed in various syndromes and disorders and how these can affect development and function within their respective field. Additionally, when a team meets in an interdisciplinary approach, mutual discussion, recommendations and consensus can be made. Here, the team is able to share ideas and opinions with one another, greatly reducing any misunderstandings or miscommunications between professions as may occur when the parents are expected to be the liaison between providers. Because the case of a child with a craniofacial disorder may be complex, it is vital that specialties work in collaboration with one another to prioritize medical and developmental needs of the child. Decisions on surgical or rehabilitative approaches can be made when the team is able to confer together and work with the goals of the parents. Austin et al. (2010) assessed perceived parental satisfaction of interdisciplinary craniofacial teams when compared to individual providers. Their survey concluded that parents of children without a team care scored medical care lower than those whose children were followed by a team of providers.

A craniofacial team is comprised of a panel of professionals who are skilled at understanding the complex needs of this population. While the number of team members can vary depending on the setting and the demands of the clinic, most craniofacial teams have a similar group of professionals present. A pediatrician is on the team to assess and monitor the general development and overall health of the child. The pediatrician may work closely with a craniofacial nurse who also assists in monitoring the overall health of the child, provides parental information on proper nutrition and feeding techniques, if needed. A geneticist provides information and assessment of possible genetic causes of the disorder, the presence of a syndrome and provides counseling on the family's

likelihood of abnormalities arising in subsequent pregnancies. Due to the nature of craniofacial abnormalities, a pediatric plastic or facial surgeon is often part of the team. The surgeon assesses the need or future need for reconstructive surgery.

In addition to these medical professions, other healthcare providers play integral roles in the team (Colburn & Sherry, 1985; Strauss, 1998). A pediatric dentist monitors the oral health and development of the child and may recommend additional consultation by an orthodontist for alignment of the teeth and jaw or an oral surgeon, for possible surgical intervention of oral structures. A psychologist is also commonly included for assessment and support of cognitive and emotional development of both the child and other family members. The psychologist can provide the family with resources and guidance on adjustment and behavioral management, if needed. A social worker is also commonly found to assist with service management and program access.

The professions of speech-language pathology, otolaryngology and audiology are also typically found on a craniofacial team. These professions work closely with one another to assess the growth and development of the auditory and oral structures and their function. Speech-language pathologists evaluate and provide recommendations on the expressive and receptive speech and language capabilities of the child as well as collaborate with the nurse and pediatrician for feeding management and therapies when needed. The otolaryngologist assesses the structure of the ear, nose and throat and identifies abnormal function in breathing, feeding and in sound conduction through the auditory pathway. The otolaryngologist also monitors and provides medical intervention for middle ear pathology, as is commonly found with this population (Di Francesco et al., 2008).

Finally, an audiologist is an important member of the craniofacial team. Audiologists work closely with the otolaryngologist and the speech-language pathologist to assess auditory function in these children. Due to the increased prevalence of both sensorineural and conductive hearing losses in this population, extensive assessment and monitoring of these children's auditory function is crucial. For children who exhibit hearing loss, the audiologist works with the child and the family to provide appropriate management of the hearing loss.

Early Intervention:

Early identification and early intervention of hearing loss is a profession-wide goal of audiology. Research has shown that better outcomes in speech and language development are closely linked to the age of identification and intervention of a hearing loss in children (Yoshinaga-Itano et al., 1998). For children with known risk factors at birth, including craniofacial abnormalities, it is vitally important that hearing is evaluated as soon as possible. Additionally, these children should be periodically monitored for changes in hearing as fluctuating thresholds due to middle ear pathology are commonly found in this population.

Universal newborn hearing screenings (UNHS) have become a widely used clinical tool for early identification of congenital hearing loss. Prior to mandated newborn screening, the average age of identification of a hearing loss was between 13-36 months (Harrison & Roush, 1996). While UNHS has been beneficial in the diagnosis of hearing loss present during the child's stay in the hospital, it may create misleading information to the parents. For the population with craniofacial abnormalities, fluctuating or late-onset hearing losses, both sensorineural and conductive, may present

after the child has passed the newborn screening. Therefore, it is important for the audiologist to be on the craniofacial team to address auditory concerns, provide periodic assessments of the child's auditory system and to begin intervention when appropriate.

As noted in research by various professions, the population of children born with craniofacial abnormalities exhibits a greater incidence of conductive hearing loss than typically developing children (AAP & ACOG, 1997; Di Francesco, Paulucci, Nery, & Bento, 2008). Due to this aspect of craniofacial abnormalities, early identification and appropriate follow-up is vitally important. Studies have shown that a child who experiences fluctuations in hearing due to middle ear effusions is at a greater risk of speech and language delay (Priester & Goorhuis-Brouwer, 2008; Sininger, Doyle, & Moore, 1999). Due to conductive hearing losses caused by middle ear effusion, these children are not receiving a steady and consistent input of speech and language. This puts them at a significant disadvantage to the incidental learning of speech and language that children with no middle ear pathology are experiencing and using in the development of their own speech and language skills.

In addition to early identification and monitoring for hearing loss in this population, the audiologist is able to provide parents information on the possible influences a hearing loss can have on speech and language development, either permanent or transient in nature. It is important for parents to understand the importance of incidental learning and consistent auditory input and their direct relationship to speech and language development. The audiologist can be a useful resource for information on enhancing auditory learning, environmental modifications to maximize hearing and listening and on possible warning signs of a change in hearing.

Diagnostic Testing

A main objective of a craniofacial team is to conduct diagnostic testing on various aspects of the child's development. For the team audiologist, this means conducting a comprehensive audiologic evaluation that is most appropriate for each child. These tests should involve both behavioral and electrophysiologic tests to assess both the integrity of the auditory system as well as the behavioral abilities of the child. Information gathered from audiologic testing can be useful in group decision making regarding both short and long term management of various aspects of development.

As the cornerstone of audiologic testing, it is important for the audiologist to obtain behavioral information on ear-specific tonal and speech stimuli in the sound booth. This information can help assess the integrity of the entire auditory system and can give insight into how the child perceives various inputs. Behavioral information obtained should include speech recognition or detection thresholds and frequency-specific responses either in the soundfield or under headphones. Due to the high prevalence of conductive and mixed hearing loss in the craniofacial population, bone conducted tonal or speech recognition information can help identify the type of hearing loss. For older children with more developed vocabulary and speech production, word discrimination abilities should also be obtained. Behavioral information is very important when determining function of the auditory system as it can be used in close comparison to objective test results in the cross check manner as described by Jerger and Hayes (1976).

In addition to behavioral assessment, the audiologist will use various electrophysiologic measurements to provide information on the integrity of various areas

of the auditory system. For some children, behavioral results are not reliable or cannot be obtained. For these patients, electrophysiologic measurements may be the best way to get accurate, frequency-specific and ear-specific information on the child's auditory system. These tests results can help estimate hearing acuity and can be used to assist with amplification. However, behavioral measurements should continue to be attempted as they are the most accurate representation of hearing acuity (Maddell, 2008).

Universal Newborn Hearing Screening, or screening the hearing of infants at birth, is now mandated for most children in the U.S. and approximately 95% of infants are screened prior to discharge (NCHAM, 2008). The screening is performed either with otoacoustic emissions (OAEs) or with an Automatic Auditory Brainstem Response (AABR) test as a screening method. For a child with a craniofacial abnormality, information should be provided to the parents at this time on the prevalence of hearing loss in this population and appropriate auditory developmental milestones to informally monitor during development, as these children are at risk for hearing loss as outlined by the Joint Committee on Infant Hearing (2000).

For newborns who are referred for further assessment based on the results of hearing screening and for children who cannot provide reliable behavioral results, an audiologist will perform an Auditory Brainstem Response (ABR) to get an accurate assessment of the auditory system. This evaluation can be completed at any age however, maturational changes in the auditory system should be considered with younger children, especially premature infants (Jiang, Brosi, Li, Chen & Wilkinson, 2005). Some infants can be assessed through non-sedated, sleep deprived testing conditions. However, many children require sedation to provide for appropriate testing parameters. When a

child is referred from a newborn hearing screening, it is typically possible to obtain non-sedated evaluations until approximately 6 months of age. At this developmental age, the child is typically too active for non-sedated testing and may need to be sedated to obtain useful and complete auditory brainstem evaluations (ASHA, 2004).

For most typically developing children, risks associated with the necessary sedation are not as significant as a child with other medical needs and therefore, an ABR evaluation is feasible. However, for many children with craniofacial abnormalities, the risks of sedation increases drastically (Butler, Hayes, Hathaway & Begleiter, 2000). Therefore, it becomes a greater challenge to obtain audiologic information, as an ABR may have to be scheduled only when another procedure requires sedation. However, when this test is able to be administered, valuable information on the type and degree of hearing loss can be obtained and applied to determine appropriate amplification and other recommendations.

Tympanometric testing should also be conducted on this population as it provides insightful information on the status of the middle ear system. Tympanometric testing can help assess the patency of PE tubes, if they are present, or the function of the tympanic membrane. Tympanometric testing plots admittance of the tympanic membrane as a function of changing air pressure (Shanks & Shelton, 1991). Acoustic reflex testing can also give insightful information on both the ipsilateral and contralateral auditory pathways and the function of the stapedius muscle (Palmu, 2005). The stapedius muscle which is responsible for the acoustic reflex contracts to high intensity stimuli and can give information on the integrity of the seventh and eighth cranial nerves and structures in the central auditory nervous system (Palmu, 2005).

Additionally, the audiologist may use Otoacoustic Emissions (OAE) information to assess the function of inner ear outer hair cells. While this test provides information on the function of an important aspect of the auditory system, it does not provide threshold information and therefore cannot be used as the sole diagnostic tool. Because of the nature of reverse emission collection in the ear canal, middle ear pathology can eliminate emissions and useful test information. Because otoacoustic information must be transmitted from the outer hair cells in the inner ear through the middle ear to be detected in the ear canal, middle ear pathology can significantly affect OAE test results (Akdodgan & Ozkan, 2006). For children who are at a higher risk for middle ear pathology, such as children born with cleft lip and cleft palate, OAE information should be evaluated with the status of the middle ear system in mind (Chen, Messner & Curtin, 2008). With the known absence of middle ear pathology, OAEs are useful in assessing cochlear function and for identifying possible auditory neuropathy and therefore should be a consistent test used when assessing the pediatric population (Madden, Rutter, Hilbert, Greinwald, & Choo, 2002).

While little information is currently available on pediatric vestibular disorders, certain causes and risk factors for these disorders are known. Etiologies that have been associated with vestibular symptoms in the pediatric population includes: prenatal drug and alcohol abuse, neurologic disorders and some genetic syndromes. Additionally, chronic middle ear pathology and middle ear effusion can manifest vestibular symptoms. Golz et al. (1998) noted that vestibular symptoms in pediatric patients were linked to the presence of middle ear effusions and noted that symptoms often resolved post insertion of PE tubes. While studies such as the Golz et al. (1998) article document the presence of

imbalance in children with effusions, no one current theory of why this happens exists. Several theories, including influence of effusions on round window movement and negative pressure influence on the inner ear have been researched (Goycoolea, Muchow & Schachern, 1988). Therefore, an audiologist working with the craniofacial population should be aware of vestibular symptoms and be able to work in collaboration with pediatric neurology, physical therapy and otolaryngology to assess balance disorders. Possible workup can include electronystagmography which can evaluate ear-specific vestibular and central function. Additionally, the Dix-Hallpike maneuver, a test that assesses dizziness with specific head motions that can help diagnosis Benign Paroxysmal Positional vertigo (BPPV). Posturaography evaluations may also be informative as they evaluate a patient's use of various sensory inputs: vestibular, visual and proprioceptive alone and in combination with one another (Casselbrant, Villardo & Mandel, 2008). Finally, as part of an interdisciplinary team, global development should be assessed through a normative evaluation such as the Peabody Developmental Motor Scale (PDMS). This documented reliable and valid evaluation can compare the function of both a child's fine and gross motor development (Folio & Fewell, 1983).

As a member of a craniofacial team, the audiologist should be well-versed in various other conditions that can affect diagnostic testing and applicable intervention. Due to the syndromic nature of many craniofacial disorders, various aspects of cognition, speech and language and gross and fine motor development may be affected. It is important that the child is tested by an audiologist who understands these aspects of a child’s development and how they may influence appropriate audiologic evaluations and intervention.

For any patient seeking audiologic services, cognitive function can affect various aspects of how diagnostic testing is administered. For a child with a craniofacial abnormality, particularly those with associated syndromes, limited cognition may be a co-morbid factor. When evaluating these children, it is important to have a pediatric and experienced audiologist conducting testing to get the most specific and accurate information.

Additionally, many of the children followed by a craniofacial team have delayed speech and language skills. Abnormal facial structure, fluctuating hearing loss and cognitive delays can all lead to the delay of expressive and receptive speech and language skills. These delays can affect the comprehension of conditioned play testing and with the validity of speech recognition and word discrimination results. In addition to delays in overall speech production and comprehension, children with craniofacial abnormalities can demonstrate abnormal resonance, vocal quality and babbling patterns (Jung, 1989). Because of these commonly found speech and language delays, a pediatric audiologist working with the craniofacial population should be able to adapt testing procedures to gain the most useful information from each child.

Finally, gross and fine motor and visual development should be taken into consideration when determining proper diagnostic audiologic tests. As a child with a craniofacial abnormality may present with delayed gross and fine motor abilities, the onset of a head turn response to sound may be delayed. This could impact typical testing protocols, however behavioral information should always be attempted. Visual impairment may influence the ability to perform Visual Reinforcement Audiometry (VRA) or typical conditioning games for Conditioned Play Audiometry (CPA). Despite

these added factors to testing, the pediatric audiologist should be able to modify testing as needed to obtain as much useful information as possible (Maddell, 2008).

Management of Hearing Loss:

In addition to early identification of a hearing loss, early intervention is crucial. Much research has shown the importance of early amplification for children with hearing loss to help promote age-appropriate speech and language skills (Yoshinaga-Itano et al., 1998; Sininger et al., 1999). For a child diagnosed with a hearing loss in a craniofacial clinic, the importance of early and appropriate amplification remains a necessity. However, given the prevalence of abnormalities to the skull and auditory pathways, traditional amplification may not always be the most appropriate device. Therefore, an audiologist who is familiar with both traditional and non-traditional amplification devices should help make amplification decisions with these families.

For some children seen by a craniofacial team, traditional amplification through air conduction hearing aids are the most appropriate. For these children, physical composition of the external ear allows for traditional hearing aids and earmolds to be worn by the child. The child's hearing loss would also be appropriately amplified by the device selected by the audiologist. The hearing aid fitting should include selecting a device that is appropriate for the child's hearing loss but also one that can be reprogrammed given a future change in hearing. The device should also be compatible with an FM system for additional connectivity in the classroom, provide sufficient channels for appropriate programming by the audiologist and be adaptable to the desired acoustic input as recommended (i.e. directional or omnidirectional microphones) (AAA,

2003). Real ear measurements or other verification tests should be conducted to ensure proper amplification as outlined by Palmer and Mormer (1999).

Depending on the degree, type and configuration of the child's hearing loss, other devices may be more appropriate than traditional amplification. For some children with craniofacial abnormalities, bone-anchored hearing aids, bone-conduction hearing aids and cochlear implants may be the most audiologic and medically appropriate device. Multiple members of the craniofacial team should collaborate together to make a decision regarding the device including the parents and the child, the audiologist and the otolaryngologist. As the team member with the expertise in hearing and amplification, the audiologist can provide the family and the team with appropriate options and their advantages and disadvantages.

As discussed earlier, one commonly found attribute of craniofacial abnormalities is chronic middle ear pathology, which may lead to significant conductive hearing losses and possible draining ears. Additionally, some children seen in craniofacial clinics have abnormalities of the external ear and/or external auditory canal. For these reasons, bone conduction or bone-anchored hearing aids may be the most appropriate device for amplification. These devices, which sit on or are imbedded in the temporal bone, do not require a device to be placed in the external auditory canal. This may be appropriate for a child with atresia, a narrowing of the external ear canal, microtia, an underdeveloped pinna, or with anotia, an absent pinna. For children with bilateral conductive or mixed hearing losses and abnormalities that prevent traditional amplification, this option may be the most appropriate (Snik et al., 2005). However, due to continued growth of the skull, children are not routinely implanted until the age of 5 years. Children with additional

craniofacial factors may not be surgical candidates or may not be ready for surgical implantation until later in development. Due to these factors, it is important for the surgical pediatric otolaryngologist to be a contributing member of this decision. Additionally, it may not be appropriate to use a surgical device if the hearing loss may be medical corrected in the future. For a child with middle ear effusion, traditional amplification or a softband bone anchored hearing aid (BAHA) may be more appropriate as the device can be removed easily if hearing returns to the normal range and amplification is no longer necessary. The BAHA device is a bone conducted processor that can stimulate inner hair function through a titanium implant or through a softband headband. Implantation has been FDA approved for children over 5 years of age.

Bone anchored devices have also been proven significantly beneficial for patients with unilateral hearing losses. For a patient with single sided hearing loss, the BAHA device works as a microphone on the ear with a hearing loss and sends acoustic information through bone conduction to the better hearing cochlea. Studies have shown improvement in sound awareness from the impaired hearing side, increased abilities to process in background noise and improvement in subjective questionnaires regarding hearing ability with adults implanted (Linstrom, Silverman & Yu, 2009). This setup would be appropriate for a child with a unilateral sensorineural hearing loss. Children with Goldenhar Syndrome may be appropriate candidates as they typically experience unilateral developmental malformations (Scholtz et al., 2001).

It is the role of the audiologist to educate others on the benefits of amplifying a unilateral hearing loss. Hol, Snik, Mylanus and Cremers (2005) assessed the audiologic and subjective differences found with using a BAHA for a child with a unilateral

conductive hearing loss. They found significant improvement in sound localization, speech recognition in the presence of background noise and a subjective improvement on quality of life questionnaires. For these reasons, it is important for a child with a unilateral hearing loss to be properly aided. The audiologist on the team must educate the parents and other professionals on the difficulties of listening in the classroom and the detrimental affect any hearing loss can have on classroom understanding and educational performance.

As with any patient demonstrating a severe to profound bilateral hearing loss, cochlear implantation may be appropriate for children with craniofacial abnormalities and this degree of hearing loss. It is important for the craniofacial team to assess various aspects of the child's physical, audiological and developmental status to determine candidacy for implantation. The audiologist must counsel parents on realistic expectations of the device, which must take into account various assessments from the other professionals on the team in additional to audiologic information.

MacArdle et al. (2002) assessed four children with craniofacial syndromes who were implanted and compared them to age-matched children with no other developmental concerns. They found equal gains in receptive language skills and in detection, recognition and identification of various environmental sounds when compared to the control group. A notable difference was seen in expressive language skills, as none of the children with craniofacial abnormalities had intelligible speech at the time of the study. Despite the small number of subjects in this study, the researchers do bring up important considerations when assessing cochlear implantation candidacy for these children. As a collaborative group, the craniofacial team must make this decision based

on a comprehensive assessment of various aspects that can provide appropriate expectations for the child post implantation. Information from detailed imaging of the cochleas, vestibular system, the brain and the seventh and eighth cranial nerves can give insight into the ability of the surgeon to get a full insertion and on the ability of the nerve to send electrical information to the brain. Parental commitment to habilitation, decisions of school placements and communication approaches must be fully discussed prior to implantation. Finally, an audiologist, prior to implantation, must assess the child's use of well-fit, appropriate hearing aids.

As illustrated, for a child with a craniofacial abnormality and hearing loss, there are many options for amplification. It is important that the family has information and realistic expectations on the various forms of amplification before making a decision. Given the level of expertise on the various forms of amplification, the craniofacial team audiologist should work closely with the family and other team members to provide the most appropriate recommendations given the various aspects of the child's hearing loss and other contributing craniofacial factors.

Monitoring of Hearing Loss:

An important aspect of audiologic services for any patient is the periodic monitoring and re-assessment of hearing and hearing loss. These assessments are particularly important for children with fluctuating or conductive hearing losses, as various medical interventions and craniofacial growth changes may lead to changes in auditory thresholds. For a child with a craniofacial abnormality, monitoring of hearing is vital to ensure early detection and appropriate intervention. Many children who visit

craniofacial clinics have ongoing middle ear pathology, and therefore may have intermittent decreases in hearing.

One aspect of monitoring the audiologist must decide is the rate at which to re-evaluation hearing. For many children, visits to the craniofacial clinic occur every 6 months in the first years of life and then increase to every year as the child reaches school age. For those patients with conductive and fluctuating hearing losses, this may be too infrequent and may need to be seen by audiology on a more frequent basis. It is the role of the audiologist to determine the rate at which a child is monitored.

If the child has a known hearing loss and amplification device, follow-up is also important as any noted change in hearing should be documented and used to adjust devices to provide the most appropriate gain. As the child grows, the amount of amplification delivered to the tympanic membrane will vary, necessitating the need for periodic real ear or functional gain measurements to verify appropriate settings. Winter and Eisenberg (1999) discuss the need for period monitoring of both audibility through real ear measurements and through behavioral response in the soundfield.

For children with purely conductive hearing losses, monitoring should coincide with medical interventions, measuring both pre and post intervention thresholds to assess changes. Additionally, the audiologist on the craniofacial team will be able to counsel the parents or caregivers on appropriate auditory developmental skills for monitoring at home. Parents should be proactive observers of the child and pursue re-evaluations if they feel the child's responses to sound have changed or if auditory developmental milestones are not being met.

It is important for the child with a craniofacial abnormality to have their hearing abilities closely monitor, given either a known hearing loss or normal hearing. The audiologist working with the craniofacial team should provide appropriate recommendations for the rate of monitoring. This recommendation will be based on a variety of factors: the presence of a hearing loss, presence of middle ear pathology, the child's age and ability to self-report a decrease in hearing and any medical intervention.

CHAPER IV

CONCLUSIONS:

As illustrated in this paper, a child born with a craniofacial abnormality will undergo significant diagnostic testing, medical procedures and rehabilitative interventions to assist with various aspects of typical development that may be adversely affected. Because of this, many parents or caregivers choose to have their child be seen by a craniofacial clinic, comprised of a panel of medical and rehabilitative experts who work in an interdisciplinary team approach. The team meets on a regular basis with the child and the family to review medical evaluations and recommendations and to make cohesive decisions on the patient's care and medical management.

Along with other head and neck abnormalities, anatomical differences and auditory dysfunction are found at a higher prevalence rate in this population when compared to typically developing children. Abnormalities have been documented in all parts of the auditory system: outer ear, middle ear, inner ear and in the central auditory nervous system. Due to these abnormalities, sensorineural, conductive and mixed hearing losses are more commonly found in this population than in children with no craniofacial abnormality. In addition to hearing loss based on anatomical and physiologic differences, middle ear pathology is also found at a higher prevalence.

For these reasons, an audiologist is a commonly found professional on the craniofacial team. The audiologist can provide a variety of diagnostic tests that can help assess the integrity of the auditory system and the hearing capabilities of the children who visit the interdisciplinary team. When a child is diagnosed with a hearing loss, the audiologist is able to provide rehabilitative recommendations to the team and to the

family. Additionally, the audiologist will closely monitor the hearing loss of the child, especially pre and post medical intervention for conductive hearing losses. In conclusion, the audiologist is able to work in collaboration with all members of the craniofacial interdisciplinary team to provide the most appropriate service that fit best with the goals and wishes of the child and the family.

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